Cystic Fibrosis
(March 2007)

Problem: Cystic fibrosis (CF) is an autosomal recessive genetic disorder of the exocrine glands, affecting approximately 30,000 children and adults in the Untied States. [(1)] Diagnosis usually occurs by six months of age. With improved treatment and earlier diagnosis, the median survival age has increased to 33.4 years. [(1)] Respiratory disease is the most recognized symptom of CF and is the primary cause for morbidity and mortality. Respiratory cultures of *Psuedomonas aeruginosa* and some strains of *Buckholderia cepacia* may suggest a poor outcome. Students diagnosed with CF are encouraged to avoid other children with CF due to possible cross infection. Social distancing of three feet is recommended for children with CF from other CF children.

Pancreatic insufficiency and damage result from obstruction of the pancreatic duct by thick secretions. Nutritional challenges exist with CF. New enzyme medications increase absorption. Compliance can be an issue. Another complication of CF can be diabetes (in second or third decade of life). The onset may or may not be symptomatic.

Role of School/School Nurse

- Communication with family is key. Developing the relationship with the family to support the student during the school day is vital.
- Complete health history should be taken including current status and recent exacerbations.
- Determine level of self-care the student has attained.
- Work with the health team (include teacher, administrator, parent/guardian, primary health care provider, CF clinic provider, student [age appropriate] and others with a vested interest) to create an individualized health care plan (IHP).
- Create a resource list of health care providers
- Care plan should include:
    - Respiratory care at school i.e. medication before meals, physical therapy, etc.,
    - Nutrition i.e. supplements before meals, caloric supplements, food diary, other monitoring tools,
    - medication compliance,
    - self-esteem (including body image),
    - risk of infection,
    - absenteeism (establish an alternative education plan for extended absences).
- Care plan must be shared with staff as needed. Trainings to care of CF child can be coordinated with the CF Foundation and parents.

Resources:

Cystic Fibrosis Foundation 6931 Arlington Road Bethesda, MD 20814 1-800-344-4823.
http://www.cff.org/home/
Cystic Fibrosis Worldwide http://www.cfww.org
Local hospitals with Cystic Fibrosis Clinics (name from Pat P.)

Reference: 1. Silkworth CK, Arnold MJ, Harrigan JF, Zaiger DS. Individualized healthcare plans for the school nurse. Sunrise Press 2005. Chapter 33 Cystic Fibrosis. 401—415.